

Anencephaly / by J.F. Knott.

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Publication/Creation

Dublin : Printed for the author by John Falconer, 1883.

Persistent URL

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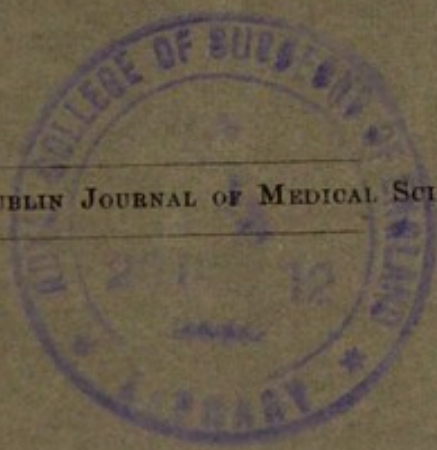
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Reprinted from the "DUBLIN JOURNAL OF MEDICAL SCIENCE"—May, 1883.

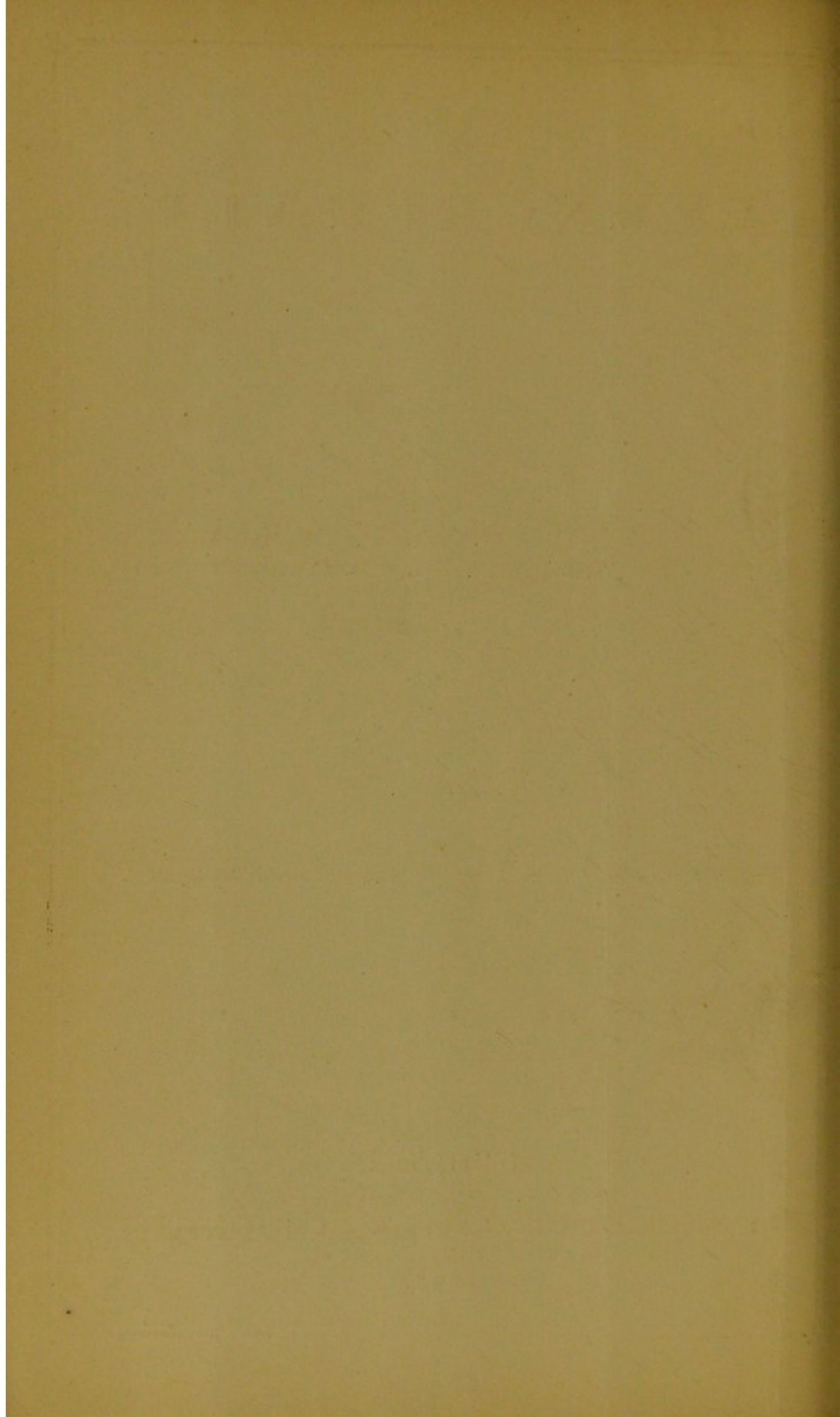


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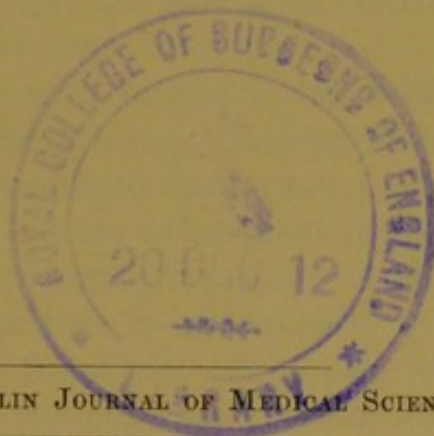


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ANENCEPHALY.*

CASES of the form of monstrosity known as anencephaly have hitherto been divided into a large number of groups, of which the characteristics are not clearly defined, because the transitional limits cannot in all cases be determined. A condition common to all these monsters is the displacement or absence of the brain, or of some of its parts, while the bones which encase the organ, or should have encased it, are more or less disarranged. Anencephalous fœtuses (excepting cases of partial encephalocele) have no chance of prolonging existence, but die soon after birth; or live, at most, only a few hours or a few days.

Anencephaly is very rare in the lower animals, as it also is indeed in case of the human being. Among 10,683 fœtuses born in the maternity of Copenhagen during ten years, there occurred but nine examples. It is twice as frequent in the female as in the male. If we wish to investigate the causes of this monstrosity, it is necessary to remember that it undoubtedly may take place at very different epochs of fœtal life. It is only when it begins to develop very early, and is complicated by the existence of spinal fissure, that it can be regarded as an arrest of development of the cranium. If, on the other hand, it supervenes at a later period of fœtal life, which, according to the researches of Professor Hannover, is usually the case, the cause is decidedly pathological, and should be sought for in the dropsy which takes place at a period more or less early in the cavities of the brain or spinal cord, and which, acting from within outwards as a mechanical force, determines a rupture and destruction of the brain substance, and prevents the formation of osseous tissue. As a result of this rupture, the galea aponeurotica and the dura mater become detached from the osseous plates already formed, from which it results not only that the bones are deprived of the basis necessary

* An abstract of Professor Hannover's Memoir—Anencephalia og Misdannelsens Forhold til Hjerneskillens Primordialbrusk.

for their future growth, but that the osseous mass already formed perishes for want of nutrition. This is the reason that the bones of the cranial vault become absorbed, while those of the base of the skull remain.

Anencephaly, if we consider man alone, commences not at the date of the origin of the hydrocephalus, but takes place when the solution of continuity occurs. The date of ossification should be borne in mind, remembering that, as has been shown in Professor Hannover's memoir on the primordial cartilage, all the bones formed between membranes, excepting the lachrymal and the lamina papyracea of the ethmoid, are ossified in the fœtus of three and a half months. If we add to this the observations made of the development of the bones during the growth of the fœtus, the rupture ought, as a general rule, to take place a month, or at most two months, after the commencement of pregnancy, when ossification is but little advanced, and the bones and their surrounding tissues are so soft and flexible that mechanical pressure can have the effect of displacing them from their normal position and modifying their form. What, nevertheless, seems to indicate that anencephalus supervenes at a later period of utero-gestation, is the age of these monsters at birth. Among the large number of cases, of which some have been observed by Professor Hannover himself, and some described by other authors, there were but few occurring between the ages of five and six months; those of seven months make a little more than a quarter, those of eight months a little over a sixth, while nearly half occurred in fœtuses which had arrived at the full term, and some were believed to have even gone beyond the period of nine months. In reality, the proportion in which the fœtus had arrived at the full term must be still greater, if we remember that these are more difficult and more expensive to preserve in our museums than specimens of less advanced development, on account of which fact they are sometimes altogether absent from collections. Examples of anencephalous fœtuses of one to two months are extremely rare.

Anencephaly, in its simplest form, engages but the bones which are formed between membranes—the vertical plate of the frontal, the parietal and squamous portions of temporal and of occipital bones; but it sometimes extends also to other bones of the cranium, when the primordial cartilage of the spine presents, at the same time, a vice of conformation in the way of a spinal fissure.

ANENCEPHALIA SIMPLEX.

The solution of continuity of the brain and its membranes usually takes place opposite the posterior fontanelle, and is found at other points only in some rare cases of partial encephalocele. Among the crania examined by Professor Hannover, there was but one found (in the Musée Dupuytren at Paris) where the rupture appeared to have taken place across the anterior fontanelle. So long as the opening is not increased, the cranium may preserve its vaulted form, especially in the anterior portion, but the vertical part of the frontal assumes a position more and more oblique, in such a way that the superciliary ridge is carried backward, and the orbit is turned more in an upward direction. The opening of the posterior fontanelle next becomes enlarged, and the surrounding bones are absorbed. The process of absorption follows a fixed rule, and takes place from behind forwards, and on the sides, but not backwards from the seat of origin of the deformity. The vertical plate of the frontal bone undergoes a gradual process of absorption, till the superciliary ridge only remains, while the horizontal portion of the bone remains. The absorption of the parietal bone becomes so complete that there remains but a thin scale lying along the superior border of the squamous portion of temporal. The latter is soon arrested in its development, and is placed a little lower than in the normal state of things, but its superior border has merely commenced to be absorbed when the parietal is found to have disappeared. With regard to the occipital bone, the superior part of the squama remains, its upper border becomes thickened and curved backwards, and it may remain in this condition even after the absorption of the parietals and of the frontal has attained its maximum; but it is also finally absorbed, and there remains only the inferior segment, which is not ossified between membranes, but is formed in what Professor Hannover has called the occipito-mastoidean portion of the primordial cartilage. The occipito-spinal membrane remains equally intact. When absorption has attained its extreme limit, a triangular opening is seen, at the bottom of which the bones of the basis cranii are exposed; these are not absorbed, but, not meeting with further resistance, are displaced towards the skin. The body and lesser wings of the sphenoid bone are prominent; they form two loops beneath which the optic foramen is seen, with its shrunken orifice transmitting

the atrophied optic nerve. In a more extreme degree of anencephaly the petrous bone lies somewhat transversely; but all the changes which supervene in that bone and in the occipital are more strongly marked in the following variety:—

ANENCEPHALIA CUM SPINA BIFIDA.

This form is scarcely as frequent as the other by one-half. While simple anencephaly is usually determined by a pathological state, the form which we have now to consider is due to a cause entirely different—that is, to an arrest of development affecting the posterior arches of the cervical vertebræ. It is difficult to decide whether that vice of conformation is due to a deficiency of cartilage, at the origin, in the primordial skeleton of the dorsal region; or if, as in the brain, it should be attributed to a dropsy which has prevented the vertebral arches from closing behind. There are cases where the serous fluid has extended without obstruction from the brain into the spinal cord; the inferior part of the squama occipitis, which represents a vertebral arch, presenting a deficiency simultaneous with that of the arches of the cervical vertebræ. But there are other cases in which that part of the occipital bone is preserved, forming a wall of separation between the pathological condition existing in the cranium and the arrest of development of the dorsal spine. This circumstance seems to indicate, in some cases, a certain degree of independence between the lesions affecting the two portions of the skeleton; but the more likely explanation is that, in most instances, the incomplete formation of the cartilage is first displayed simultaneously with the hydrorachis, and that the collection of serous fluid comes from the brain.

On examination of the bones of intra-membranous origin, we find that this form of anencephaly does not differ in any essential particular from the one previously described, but in the present variety the monstrosity reaches its greatest dimensions, the extent of osseous absorption being very great, the whole of the vertical portion of the frontal may be absent, or represented by the superciliary arches only, and the horizontal plate is narrower from before backwards than in the ordinary state. The parietal bone is reduced to a thin scale, which, by its anterior extremity, rests on the external orbital process of the frontal, and, by its posterior extremity, is united with the ossified portion of the occipito-mastoid segment of the primordial cartilage. The squama temporis is nar-

rowed and placed low, and may be so much affected by pressure as to have its external surface turned downwards; the membrana tympani may be horizontal in position, and carried towards the middle line of the body. The complete rotation of the os petrosum, described by Professor Hannover in his memoir on the primordial cartilage, does not seem to take place in anencephalic subjects.

On the other hand, the bones which are developed in the primordial cartilage undergo great changes, both in form and position. While the superior segment of the squama occipitis, which is ossified in membrane, is always deficient in these cases, we may find traces of the inferior segment, which is ossified in the occipito-mastoid portion of the primordial cartilage; when the spina bifida is more pronounced, there is also a fissure of the squama, and the two lateral portions are pushed outwards, and placed obliquely with regard to the longitudinal axis of the body. The union of each with the scale to which the parietal bone is reduced continues without change when the position of the former becomes gradually horizontal, in such a way that the face normally turned towards the interior comes to look directly upwards, and the posterior surface vertically downwards. When the spinal fissure is of still greater extent, the two portions of the squama come at last to assume a position completely vertical, in such a way that the surface originally anterior and central is now turned outwards, and the surface originally turned backwards is directed towards the middle line of the body. This characteristic change, by which the two portions of the squama occipitis are so completely folded down, is especially due to the contraction of those muscles of the back of the neck and occipital region which are attached to the outer surface of the affected bone, and whose action, in the absence of the vault of the cranium, there is no resistance to overcome. From this comes the singular aspect of the anencephalic fœtus—it seems to have no neck, while the head rests upon the back, which indeed is partly the case, the malformed external ear also touching the shoulder. The scale of the parietal bone has also been folded down on the side of the neck, where it must be searched for. Between the posterior extremity of the occipito-mastoid portion (*i.e.*, the inferior part of the squama occipitis) and the free extremity of the open posterior arch of the atlas, a cartilaginous connexion sometimes forms, which must not, however, be confounded with another connexion that sometimes takes place, in the normal

state, between the jugular apophysis and the transverse process of the atlas (Cruveilhier).

The condyloid part of the occipital bone is drawn more and more outwards and downwards by the occipito-mastoid segment, then becomes longer and narrower, and takes a position, at first oblique, then nearly transverse. The semilunar ossification, behind the condyloid portion of the bone, is carried forwards, and so is the petrous bone, which comes to occupy a transverse position, pushing before it the great wing of the sphenoid. The internal auditory meatus is turned so as to look almost directly upwards, while the deformed fossa subarcuata is directed outwards. The osseous tubercle formed by the inferior and external semicircular canals, after projecting through the primordial cartilage, is similarly displaced outwards and forwards; the canals become deformed, and hearing is destroyed as well as sight. The body of the sphenoid undergoes compression, the vertical plate of the sella turcica (*dorsum sellæ*) is turned forwards to a flattened position, the lesser wings of the sphenoid form two loops, and the optic foramen is depressed. As Professor Hannover has already pointed out, in his memoir on the primordial cartilage, the hypothesis of the basilar portion of the occipital bone being the double body of a vertebra with two arches (analogous to the atlas with the axis and its double body), seems to be specially confirmed by examination of the anencephalous foetus in which *spina bifida* co-exists.



